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The Levoatriocardinal Vein: Morphology and Echocardiographic Identification of the Pulmonary–Systemic Connection

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Objectives. **This study considers the array of pulmonary–systemic connections made by the levoatriocardinal vein. The primary and associated lesions that play a role in forming this vein are examined, and echocardiography is discussed as a method for its rapid identification.**

Background. **The levoatriocardinal vein is a pulmonary–systemic connection that provides an alternative egress for pulmonary venous blood in left-sided obstructive lesions. It is thought to result from the persistence of anastomotic channels that connect the capillary plexus of the embryonic foregut to the cardinal veins. Only 12 cases of levoatriocardinal vein have been reported since its first description in 1926. A comprehensive description of the morphology and echocardiographic identification of this lesion has been unavailable because of its rarity.**

Methods. **A retrospective study was performed in 13 patients with a levoatriocardinal vein from the University of California, San Francisco. Echocardiographic findings were compared with those obtained by angiography or at necropsy. In addition, the details of 12 previously published case reports were reviewed. Age at presentation, primary obstruction to pulmonary venous return, integrity of the atrial septum and origin and drainage of the levoatriocardinal vein were compared.**

Results. **Patient age at presentation was <2 years, with most patients presenting before age 6 months. Variations of the hypoplastic left heart syndrome accounted for the majority of primary defects encountered, although multiple but less severe left-sided lesions were seen. The atrial septum was functionally intact in most patients. The levoatriocardinal vein, defined echocardiographically, originated predominantly from the smooth-walled left atrium and drained to the superior vena cava or innominate vein; however, variations of this pattern existed.**

Conclusions. **As a physiologic entity, the levoatriocardinal vein provides a mechanism for decompression of pulmonary venous return primarily in patients with left ventricular inflow obstruction. A levoatriocardinal vein is thought to form when the atrial septum fails to provide an alternate egress for left atrial blood. However, when a septal defect or alternative shunt occurs in conjunction with a levoatriocardinal vein, the clinical presentation may be postponed. Echocardiography provides a rapid, noninvasive modality for identifying the pulmonary–systemic connection, which may masquerade as the vertical vein in anomalous pulmonary venous connection or act as an occult source of left to right shunting in patients undergoing surgery for hypoplastic left heart syndrome.**

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In most cases of severe left-sided obstruction, pulmonary venous blood passes from the left atrium through an interatrial communication to the right atrium (1). When the fossa ovalis becomes restrictive, another route may exist to decompress the obstruction of pulmonary venous return to the heart. The levoatriocardinal vein, a pulmonary–systemic connection first reported by McIntosh in 1926 (2), provides such an alternate egress for pulmonary venous blood. To our knowledge, a total of 12 patients with levoatriocardinal vein have been reported since its first description (2–11). However, because of the rarity of this vein, a comprehensive description of it has been unavailable.

During the past 14 years we have identified 13 additional patients at our institution with echocardiographic or angiographic evidence of a levoatriocardinal vein. In this retrospective study we describe the spectrum of pulmonary-systemic connections seen in these 25 patients and place these observations in the context of cardiovascular development. We examine the relative frequency of the primary obstructive lesions and associated cardiac defects and the role of atrial septal integrity in formation of the levoatriocardinal vein. The usefulness of echocardiography in detecting this vascular connection is discussed.

Methods

Study patients. All 13 patients who presented to the University of California, San Francisco from 1980 to 1994 and

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were found to have a levoatriocardinal vein were included in this study. Details of their clinical presentation and physical examination were obtained from medical records. Anatomic information was obtained by retrospective review of echocardiograms or cineangiograms or by examination of pathologic specimens. In addition, the case reports of 12 patients published between 1926 and 1993 were reviewed.

Anatomic features examined. The primary obstructive lesion, any associated cardiac defects and the state of the atrial septum were determined for each patient. We defined the levoatriocardinal vein as an abnormal venous connection that allowed return of pulmonary venous blood to the right atrium in the presence of otherwise normally connected pulmonary veins. The precise origin and drainage of the levoatriocardinal vein in each patient seen at the University of California, San Francisco was demonstrated from the two-dimensional echocardiogram complemented by color flow, pulsed and continuous wave Doppler ultrasound, or from a cineangiogram, or both.

The conduct of this study conformed to the guidelines of the Committee for Human Research at the University of California, San Francisco for performing a retrospective review of clinical material.

Statistics. To demonstrate that both groups of patients were representative of the population of patients likely to have a levoatriocardinal vein, a two-tailed test of significance using the Student *t* distribution was performed to compare the mean ages at the time of presentation between the group seen at the University of California, San Francisco and those patients reported on previously.

Results

The clinical and anatomic findings obtained for the 13 patients who presented to the University of California, San Francisco, as well as those reported for the 12 patients since 1926 are presented in Table 1.

Clinical demographics. The age at presentation for congenital obstruction to left ventricular inflow is related to the degree of obstruction, the availability of an alternate egress for pulmonary venous return and the resulting pulmonary edema. We therefore compared the age at presentation between the two patient groups to confirm that both samples were representative of the population of patients likely to have a levoatriocardinal vein. For the 13 patients who presented to the University of California, San Francisco with a levoatriocardinal vein, 12 had coincident left-sided obstruction. The age range at the time of presentation for these patients was 12 h to 19 months (mean ± SD 2.4 ± 5.6 months). This range was not significantly different ($p > 0.1$) from that of the 12 previously described patients, whose age at presentation ranged from 30 min to 22 months (mean 3 ± 6.4 months). The patients' clinical presentation uniformly reflected the decreased cardiac output and pulmonary venous obstruction caused by the primary obstructive lesions. One patient seen at the University of California, San Francisco (Patient 25) did not have a left-sided obstructive lesion and incidentally was found to have a levoatriocardinal vein by angiography at age 15 years.

Primary obstruction. Variations of the hypoplastic left heart syndrome (severe stenosis or atresia of the aortic or mitral valve, or both) accounted for the primary obstruction in 19 patients (76%); cor triatriatum was found in 4 (16%). Two patients (8%) had no obstruction to left ventricular inflow. In one of these two (Patient 24), a juxtaductal coarctation of the descending aorta was found to be the only obstruction to left-sided outflow. In the other (Patient 25), an atrial septal defect initially was suspected on clinical examination, and he was referred for cardiac catheterization after M-mode echocardiography available at the time could not confirm the diagnosis. Ultimately, he was found to have a levoatriocardinal vein but otherwise normal intracardiac anatomy by cineangiography.

Of the patients with hypoplastic left heart syndrome, one (Patient 10, from our institution) survived without intervention until age 13 years. Her initial presentation and survival through 7 years have been reported elsewhere (12). The pathologic specimen shown in Figure 1 details the presence of a levoatriocardinal vein, which may have accounted in part for her prolonged survival.

Integrity of interatrial septum. The atrial septum was intact in 14 patients (56%); an additional 7 patients (28%) had a hemodynamically insignificant patent foramen ovale. These data are consistent with the hypothesis that the levoatriocardinal vein persists in the absence of an atrial septal defect to decompress the pulmonary venous circulation.

Pulmonary–systemic venous connection. The levoatriocardinal vein is easily demonstrated by angiography (Fig. 2). In our experience, its course was best appreciated echocardiographically from the subcostal (Fig. 3 and 4) and suprasternal notch (Fig. 5) views. The spectrum of connections seen among the 25 patients is listed in Table 1. The levoatriocardinal vein originated directly from the left atrium in eight of the patients seen at our institution (Fig. 3) and in nine of the previously described patients (68%). In the remaining eight patients (32%), the levoatriocardinal vein arose from one of the pulmonary veins; however, in each of these cases the pulmonary vein continued its course to the heart distal to the origin of the levoatriocardinal vein.

Drainage of the levoatriocardinal vein was more variable. In nine patients (36%), the vein drained directly to a right-sided superior vena cava (Fig. 4), whereas in 12 patients (48%) the levoatriocardinal vein was observed connecting to the left innominate vein. The levoatriocardinal vein drained to one of the jugular veins in two patients (8%) and to a left-sided superior vena cava in one patient (4%) from our institution with left atrial isomerism and an interrupted inferior vena cava with azygos continuation. For one of the previously described patients, the drainage of the levoatriocardinal vein could not be determined because the vessel was severed from its systemic venous connection at necropsy (8).

Table 1. Clinical and Anatomic Features of 25 Patients With a Levoatriocardinal Vein

Pt No.	Age at Presentation/ Gender	Primary Obstruction	Associated Cardiac Defects	Atrial Septum	Levoatriocardinal Vein		Mode of Diagnosis*	Reference No.
					Origin	Drainage		
1	8 days/F	MA		PFO	RUPV	IV	Angio	UCSF
2	30 min/F	MA		Intact	LA	RSVC	Necro	(3)
3	1 wk/M	MA	VSD	Intact	LUPV	IV	Necro	(4)
4	1 wk/M	MA	PA/AVS	Sec ASD	LA	IV	Angio	(5)
5	2 wk/M	MA		PFO	LA	IV	Angio	(6)
6	3 mo/M	MA	Unroofed CS	Intact	LA	IV	Necro	(7)
7	4 mo/NR	MA	DORV	Intact	LA	†	Necro	(8)
8	12 h/F	MA, AoA		PFO	LA	RSVC	Echo	UCSF
9	1 day/F	MA, AoA		Intact	RUPV	RSVC	Echo	UCSF
10	1 h/F‡	MA, AoA	Aberrant left SCA, persistent DA	Intact	LA	IV	Echo	UCSF
11	2 days/M	MA, AoA	Unroofed CS	Intact	LA	IV	Echo	UCSF
12	3 wk/F	MA, Hypo arch	Multiple muscular VSDs	Intact	LA	Left IJV/SCV junction	Necro	(9)
13	5 wk/M	MA, Hypo arch	AVS	Intact	LA	RSVC	Necro	(2)
14	12 h/M	MA, Coarct	Left atrial isomerism, LSVC, interrupted IVC, VSD	Intact	LA	LSVC	Angio	UCSF
15	11 wk/F	MA, Coarct	VSD	Intact	LUPV	IV	Angio	(10)
16	12 h/M	AoA, MS		Intact	RUPV	RSVC	Echo	(3)
17	1 day/F	MS	DORV, PS	Intact	LA	IV	Echo	UCSF
18	19 mo/M	MS	DORV, PS	Multiple ASDs	LA	JV	Echo	UCSF
19	11 days/M	MS, AS, Coarct		Intact	RUPV	RSVC	Angio	UCSF
20	4 wk/M	Cor triatriatum		PFO	LA	RSVC	Echo	UCSF
21	10 wk/F	Cor triatriatum		PFO	LUPV	IV	Echo	UCSF
22	3 mo/F	Cor triatriatum		PFO	LA	RSVC	Echo	UCSF
23	3 wk/F	Cor triatriatum, Coarct		Sec ASD	LA	IV	Necro	(3)
24	22 mo/M	Coarct	d-TGA, VSD, CS drainage to LSVC	Sec ASD	LA/LAA junction	Right IJV	Necro	(11)
25	15 yr/M	None		PFO	LUPV	RSVC	Angio	UCSF

*For Patients 18 and 19, identification of a levoatriocardinal vein was made by cineangiography only, although two-dimensional echocardiography was performed; two-dimensional echocardiography was not available for Patients 1 and 25. †Drainage of the levoatriocardinal vein could not be determined because the vessel was severed from its systemic venous connection at necropsy. ‡Survived to age 13 years without intervention; her survival was most likely facilitated by the coexistence of a levoatriocardinal vein with a persistent ductus arteriosus (DA), which served to decompress the obstruction to left ventricular inflow. Angio = angiogram; AoA = aortic atresia; AS = aortic stenosis; ASD = atrial septal defect; AVS = absent ventricular septum; Coarct = coarctation of the aorta; CS = coronary sinus; DORV = double-outlet right ventricle; d-TGA = dextrotransposition of the great arteries; Echo = echocardiogram; F = female; Hypo arch = hypoplastic aortic arch; IJV = internal jugular vein; IV = innominate vein; IVC = inferior vena cava; JV = jugular vein; LA = left atrium; LAA = left atrial appendage; LSVC = left-sided superior vena cava; LUPV = left upper pulmonary vein; M = male; MA = mitral atresia; MS = mitral stenosis; Necro = necropsy; NR = not reported; PA = pulmonary atresia; PFO = patent foramen ovale; PS = pulmonary stenosis; Pt = patient; RSVC = right-sided superior vena cava; RUPV = right upper pulmonary vein; SCA = subclavian artery; SCV = subclavian vein; Sec ASD = ostium secundum atrial septal defect; UCSF = University of California, San Francisco; VSD = ventricular septal defect.

Discussion

Development of levoatriocardinal vein. The pulmonary veins are believed to develop from a capillary plexus surrounding the embryonic foregut (9). These coalesce to form the individual main pulmonary veins that drain each lobe of the lung. Ultimately, these main pulmonary veins join with an outgrowth of the sinoatrial region of the heart to form the connection between the pulmonary venous bed and the left atrium. Early in this process, the capillary plexus of the developing esophagus has connections with vessels of the cardinal system, which eventually give rise to the innominate and jugular veins, the superior vena cava and the coronary sinus (10). Usually these connections are lost as the pulmonary bed establishes drainage to the heart, but in cases of proximal pulmonary venous obstruction it is thought that these vessels persist and form the basis for anomalous pulmonary venous connection. However, if obstruction to pulmonary venous drainage occurs within the heart, these pulmonary–systemic connections can provide for the egress of pulmonary venous blood from the left atrium.

The name "levoatrio-cardinal vein," first recommended by Edwards and DuShane (9), describes the physiologic phenomenon of left atrial decompression through the cardinal venous system. For this reason, we and others (3–5,10) believe that this term is appropriately applied regardless of whether there is a direct connection with the left atrium. However, a distinction should be made between this phenomenon and cases where part or all of the pulmonary venous return drains solely

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Figure 1. Pathologic specimen demonstrating the levoatriocardinal vein in a patient with hypoplastic left heart syndrome. **A,** Heart viewed from the front with the large right ventricle (RV) and right atrial appendage (RAA) identified. The hypoplastic aortic root (Ao) is seen, and the large pulmonary artery (PA) has been opened as it exits the heart. The entrance of the levoatriocardinal vein (**arrow,** LACV) into the innominate vein (IV) is identified, and the latter can be seen draining to the right superior vena cava (SVC), which in turn drains to the body of the right atrium. **B,** Course of the levoatriocardinal vein (**large arrow**) from the left atrium. The posterior aspect of the heart is seen, and a vestigial left ventricle (LV) in contrast to the large right ventricle can be appreciated. The left atrium (LA) has been opened, and the **three small arrows** indicate the junction of the pulmonary venous confluence, or posterior chamber, with the body of the atrium, or anterior chamber. The left (LPV) and right (RPV) pulmonary veins can be seen entering the posterior chamber. **C,** Origin of the levoatriocardinal vein from the left atrium as it ascends adjacent to the aortic root. The left lung has been reflected to the right so that the left lower pulmonary veins (LLPV) can be seen draining to the left lower corner of the left atrium. The left pulmonary artery (LPA) can be seen supplying the left lung. **D,** Close-up view of the left atrium and the draining of the left and right pulmonary veins into its posterior aspect. The left (LB) and right (RB) bronchus can be seen over the superior border of the left atrium. A small portion of the right atrial appendage can also be seen.

through the cardinal system, bypassing the heart completely. In such cases, the anatomy dictates that it be referred to as "anomalous pulmonary venous connection."

The levoatriocardinal vein also should be considered an entity distinct from a left-sided superior vena cava. The latter forms from the left precardinal vein when the left innominate vein fails to develop normally (9). The left innominate vein may be present as a small bridging vein that obliquely connects the left and right precardinal systems, or it may not form at all. As proof of this distinction, one of the patients seen at the University of California, San Francisco (Patient 14), had both a left superior vena cava and a levoatriocardinal vein that drained to it, and Harris et al. (11) described a patient (Patient 24) with a levoatriocardinal vein who also had coronary sinus drainage to a left superior vena cava.

Primary defect and role of interatrial septum. In our series, a range of left-sided obstructive lesions was encountered in association with a levoatriocardinal vein, including mitral atresia, aortic atresia, coarctation and instances of multiple levels of obstruction to left-sided outflow. One might predict that the more proximal the level of obstruction, the less opportunity for relief of the obstruction by an intracardiac left to right shunt. If patients with such lesions survive fetal life, it seems likely that an obligatory pulmonary–systemic connection would exist, in the form of a levoatriocardinal vein, to provide relief from pulmonary venous obstruction. In fact, this occurred in our series, in which the majority of patients with a levoatriocardinal vein had a proximal obstruction in the form of mitral valve atresia, stenosis or cor triatriatum.

Two patients with a levoatriocardinal vein did not have proximal pulmonary venous obstruction. One (Patient 25) was found to have a levoatriocardinal vein incidentally on angiog-

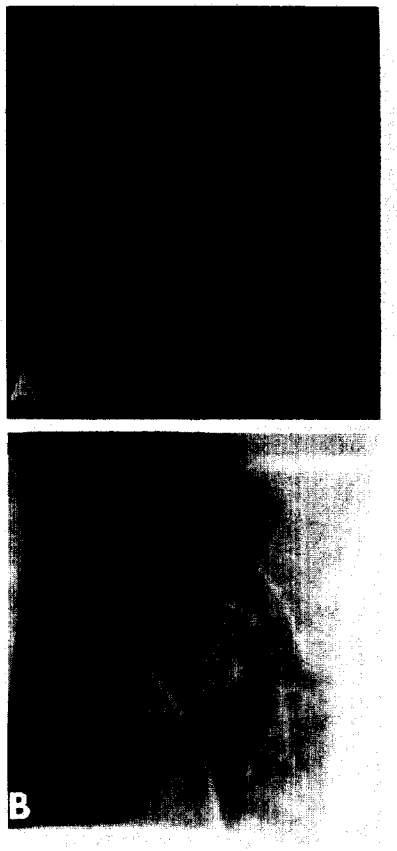

Figure 2. Angiographic appearance of the levoatriocardinal vein in a patient with hypoplastic left heart syndrome. **A,** Posteroanterior cineangiographic projection after a left pulmonary artery injection. On the recirculation phase, the pulmonary veins (PV) can be seen entering the superior aspect of the left atrium. The levoatriocardinal vein can be traced from its exit from the posterior wall of the left atrium to its junction with the prominent innominate vein. This drains to the right superior vena cava (RSVC) and superimposed right atrium (RA). **B,** Corresponding lateral cineangiographic projection of the angiogram shown in **A.** The pulmonary vein can be seen entering the superior aspect of the left atrium, but the exit of the levoatriocardinal vein is obscured by the superimposed pulmonary vein. The flow from the left atrium, through the levoatriocardinal vein, to the innominate vein, right superior vena cava and right atrium again is identified. Other abbreviations as in Figure 1.

raphy when he presented for evaluation of left to right shunt; the other (Patient 24), described by Harris et al. (11), had a coarctation as the only left-sided obstructive lesion. Perhaps in this latter case, the periductal shelf formed at the coarctation obstructed blood flow in both the aorta and the ductus arteriosus, resulting in a hemodynamic state dependent for survival on the existence of a levoatriocardinal vein.

The occurrence of a levoatriocardinal vein, with intracardiac obstruction to pulmonary venous return, usually is associated with an intact atrial septum (3,9). However, in this series of 25 patients, 4 (16%) had a hemodynamically significant atrial septal defect. This finding demonstrates that a functionally intact atrial septum is not absolutely necessary for the formation of a levoatriocardinal vein. Of these four patients with a defective atrial septum, two did not present until nearly 2 years of age, which was well beyond the mean age of presentation for this group of patients with severe left-sided obstruction. Most likely, this second level of decompression ameliorated the early hemodynamic compromise seen in the other patients. Similarly, one patient (Patient 10) seen at our institution with mitral and aortic atresia had a persistent ductus arteriosus in addition to the levoatriocardinal vein; this patient survived to 13 years of age without any intervention and is perhaps the longest living patient with unoperated hypoplastic left heart syndrome reported to date.

Pulmonary–systemic venous connection. This series of 13 patients seen at the University of California, San Francisco in combination with 12 previously reported patients provides the opportunity to describe the pulmonary-systemic connection made by the levoatriocardinal vein. We observed a wide spectrum of origin and drainage. When the connection became incorporated into the smooth-walled portion of the left atrium, the result was a direct communication between the left atrium and the cardinal venous system. This appeared to be the predominant pattern for this anatomic entity, occurring in 68% of the 25 study patients. When a more proximal connection within the lung formed, the communication developed between one of the pulmonary veins and the cardinal veins, accounting for the remaining 32% of cases. In no case did the anomalous vein originate from either the trabeculated portion of the left atrium or the precardinal system, thereby confirming its identity distinct from the left-sided superior vena cava.

The majority of pathways observed resulted in decompression of left atrial blood to either the superior vena cava or the left innominate vessel. Only a few of the levoatriocardinal veins drained to more distal vessels of the cardinal system. Although the primordial anastomoses between the capillary plexus of the foregut and the cardinal veins might be expected to exist in a more widespread fashion, only those connections with the larger, proximal cardinal vessels persist as a levoatriocardinal vein. Perhaps drainage to more distal vessels is incompatible with embryonic survival.

Role of echocardiography. Two-dimensional echocardiography and color flow Doppler mapping have been used to delineate the drainage site and presence and site of obstruction in patients with anomalous pulmonary venous connection (13). When considering the latter diagnosis, it is important to recognize that a levoatriocardinal vein may mimic the vertical vein seen in anomalous connection of the pulmonary veins above the diaphragm, and an echocardiographic search for left-sided obstructive lesions should be pursued. Conversely, the finding of left ventricular inflow obstruction should always prompt the search for a levoatriocardinal vein, as this vein may provide a potential source of left to right shunting in patients with hypoplastic left heart syndrome who are to undergo the first stage of a single-ventricle repair (14).

Multiple views are required to trace the origin, relation to the atrium and the distal course of the levoatriocardinal vein. The initial approach includes subcostal views of the left atrium in both the coronal and sagittal planes, whereby the presence of left atrial outflow obstruction is identified. The subcostal

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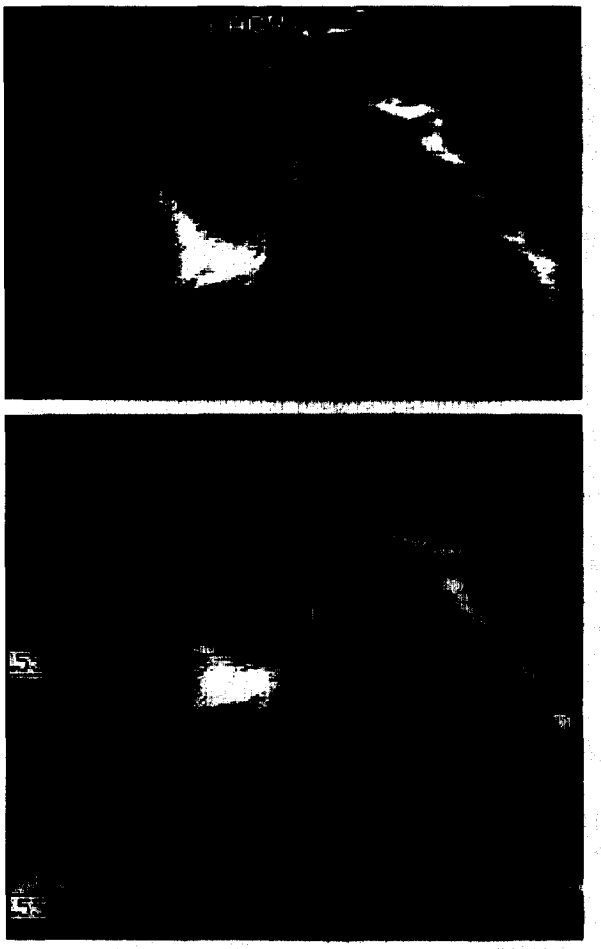

Figure 3. Echocardiographic demonstration of the levoatriocardinal vein and its origin. **A,** Subcostal coronal view. Both pulmonary veins can be seen entering the left atrium. The right and left atria are separated by an intact atrial septum. The levoatriocardinal vein can be seen exiting the pulmonary venous confluence and ascending medial to the left pulmonary artery. **B,** Color flow image of the anatomy seen in **A** identifies red flow returning from the pulmonary veins to the left atrium and blue flow through the levoatriocardinal vein to its junction with the superior vena cava (not shown). Abbreviations as in Figures 1 and 2.

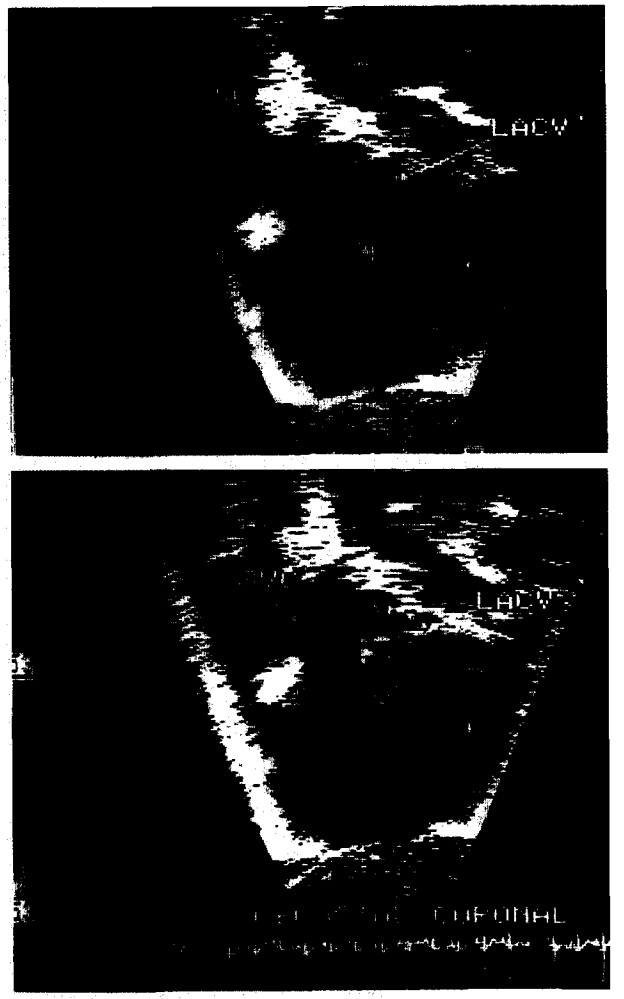

Figure 4. Echocardiographic demonstration of the levoatriocardinal vein and its drainage. **A,** Subcostal coronal view demonstrating the levoatriocardinal vein draining above the left atrium and directly into the right superior vena cava. The junction of the levoatriocardinal vein with the left atrium is not seen from this view. **B,** Superposition of color flow on the image in **A,** demonstrating the flow in the levoatriocardinal vein directly above the flow seen within the right atrium. Abbreviations as in Figures 1 and 2.

and apical four-chamber views also are used to identify the position of pulmonary veins, noting the connection of these structures with the left atrium. The venous drainage away from the atrial cavity is then interrogated. Its position and vertical ascent are traced by two-dimensional echocardiography and superimposed Doppler color flow techniques in subcostal (Fig. 3 and 4), apical, parasternal short-axis and suprasternal planes (Fig. 5). An oblique high parasternal view, parallel to the spine, may also be used to note the course and systemic connection of the levoatriocardinal vein, utilizing a technique similar to that for defining a left superior vena cava to coronary sinus connection (15). Color flow, pulsed and continuous wave Doppler ultrasound studies demonstrate the direction of blood flow. Venous flow away from the left atrium provides corroborative evidence to that of two-dimensional morphology for the presence of a levoatriocardinal vein (Fig. 5).

In our series at the University of California, San Francisco, a levoatriocardinal vein was identified in 9 of 11 patients evaluated by echocardiography. Although angiography performed in nine of the patients identified the levoatriocardinal vein in each, we cannot compare the sensitivities of these methods, as we could not control for the availability of color flow Doppler echocardiography or the experience or level of suspicion of the sonographer or angiographer in this retrospective series. However, echocardiography does provide a rapid, noninvasive modality for establishing the anatomy of the levoatriocardinal vein. This observation is important as many patients with a levoatriocardinal vein present in profound distress because of their primary obstructive lesion. Therefore, in most of these patients, clinical evaluation must be expedited to facilitate surgical intervention.

Conclusions. As a physiologic entity, the levoatriocardinal vein provides a mechanism for decompression of the

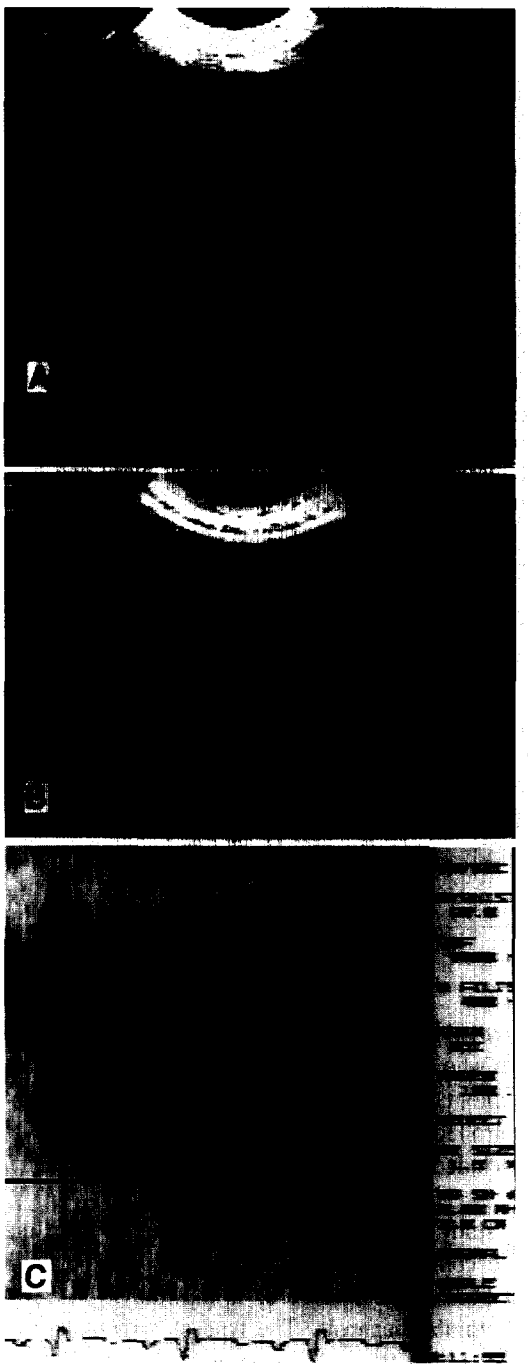

Figure 5. Use of pulsed wave Doppler echocardiography in demonstrating the levoatriocardinal vein. **A,** Suprasternal notch coronal view of the origins of the left and right innominate veins as they join to form the superior vena cava. The aorta and pulmonary artery are identified. The pulmonary veins can be seen entering the left atrium adjacent to the levoatriocardinal vein on the left and more distally on the right. The levoatriocardinal vein exits the left atrium and joins directly with the superior vena cava. **B,** Doppler reference view similar to the view in **A,** showing the sample volume placed within the levoatriocardinal vein. **C,** Pulsed wave Doppler signal coming toward the transducer with an average velocity of 70 cm/s and peak velocity with atrial contraction of almost 100 cm/s. The **scale** at right is in increments of 10 cm/s. Abbreviations as in Figures 1 and 2.

pulmonary venous circulation primarily in patients with left ventricular inflow obstruction. It is thought to form when an intact atrial septum fails to provide an alternate egress for left atrial blood. However, the occurrence of a septal defect or alternative shunt in conjunction with a levoatriocardinal vein may postpone the clinical presentation. A survey of this entity's anatomic course is consistent with its arising from the capillary plexus of the embryonic foregut to form a persistent connection with the cardinal system. Echocardiography provides a rapid, noninvasive modality for identifying the levoatriocardinal vein, which may masquerade as the vertical vein in anomalous pulmonary venous connection or act as an occult source of left to right shunting in patients undergoing surgical treatment for hypoplastic left heart syndrome.

References

1. Beckman CB, Moller JH, Edwards JR. Alternate pathways to pulmonary venous flow in left sided obstructive anomalies. Circulation 1975;52:509–16.
2. McIntosh CA. Cor biatriatum triloculare. Am Heart J 1926;1:735–44.
3. Pinto CAM, Ho SY, Redington A, Shinebourne EA, Anderson RH. Morphological features of the levoatriocardinal (or pulmonary-to-systemic collateral) vein. Pediatr Pathol 1993;13:751–61.
4. Taybi H, Kurlander GJ, Luri PR, Campbell JA. Anomalous systemic venous connection to the left atrium or to a pulmonary vein. Am J Roentgenol 1965;94:62–77.
5. Blieden LC, Schneeweiss A, Deutsch V, Neufeld HN. Anomalous venous connection from the left atrium to the cardinal venous system: "levoatriocardinal vein". Am J Roentgenol 1977;129:937–8.
6. Al-Fadley F, Galal O, Wilson N, Aloufi S. Cor triatriatum associated with total anomalous pulmonary venous drainage in the setting of mitral atresia and a restrictive interatrial communication. Pediatr Cardiol 1992;13:125–6.
7. Broustet P, Bricaud H, Mullon P, Delbancut O. Atrésie mitrale complete avec une veine cave implantée dans l'oreillette gauche assurant seule le retour du sang pulmonaire au coeur droit. Arch Mal Coeur 1959;52:1397–409.
8. Bellet S, Gouley BA. Congenital heart disease with multiple cardiac anomalies. Report of a case showing aortic atresia, fibrous scar in myocardium and embryonal sinusoidal remains. Am J Med Sci 1932;183:458–65.
9. Edwards JE, DuShane JW. Thoracic venous anomalies. Arch Pathol 1950; 49:517–37.
10. Lucas RV, Lester RG, Lillehei CW, Edwards JE. Mitral atresia with levoatriocardinal vein. A form of congenital pulmonary venous obstruction. Am J Cardiol 1962;9:607–13.
11. Harris HA, Gray SH, Whitney C. The heart of a child aged twenty-two months presenting an anomalous vein from the pulmonary auricle to the right internal jugular vein, transposition of the great vessels and left superior vena cava. Anat Rec 1927;36:31–49.
12. Ehrlich M, Bierman FZ, Ellis K, Gersony WM. Hypoplastic left heart syndrome: report of a unique survivor. J Am Coll Cardiol 1986;7:361–5.
13. Van Hare GF, Schmidt KG, Cassidy SC, Gooding CA, Silverman NH. Color Doppler flow mapping in the ultrasound diagnosis of total anomalous pulmonary venous connection. J Am Soc Echocardiogr 1988;1: 341–7.
14. Norwood WI. Hypoplastic left heart syndrome. Cardiol Clin 1989;7:372–85.
15. Silverman NH. Pediatric Echocardiography. Baltimore: Williams & Wilkins, 1993:515–6.